Prevalence of clinical characteristics of patients with a (probable) Behçet*s syndrome in the Netherlands, a follow up cohort study.

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Ethical review	Approved WMO
Status	Recruiting
Health condition type	Immune disorders NEC
Study type	Observational invasive

Summary

ID

NL-OMON45634

Source
ToetsingOnline

Brief title
Behcet study, longitudinal follow up

Condition

- Immune disorders NEC

Synonym
Behcet Syndrome, Behcet's disease

Research involving
Human

Sponsors and support

Primary sponsor: Jan van Breemen Instituut

Source(s) of monetary or material Support: Jan van Breemen research grant (voor een deel van de studie);Voor de differentiaal diagnose is er Reade reumatologie budget en voor de rest zal een aanvraag voor een grant geschreven worden

Intervention

Keyword: Behcet syndrome, immunology, observational study, questionnaires

Outcome measures

Primary outcome

No endpoints were defined, since it is an observational cohort study.

Secondary outcome

We wil record occurrence of disease manifestations and treatment given.

Disease activity and QoL will be assessed using various questionnaires.

We will measure NETosis by levels of circulating nucleosomes and systemic

neutrophil activation will be assessed by released neutrophil elastase complex

with its natural inhibitor alfa1-antitrypsine (ELA complexes) and levels of

circulating nucleosomes.

T cel regulation will be measured by clonal expansion.

Study description

Background summary

Behçet*s syndrome (BS) is characterized by orogenital ulcera and uveitis and is most common in countries along the Silk Road. It is an orphan disease in Western countries. Presentation varies with gender and also with geographical area. This implicates that classification criteria based on an endemic population might not be adequate for Western patients. At this time incomplete data on the presentation of patients with (suspected) BS in the Netherlands are present. This complicates diagnosis and also prediction of likelihood of developing BS. The exact pathogenesis remains unclear. Environmental and infectious factors are thought to play a role in the development of the disease. Furthermore, 10% of patients in Turkey have a positive family history,

suggesting a role for genetic factors. Also, BS has both histological and clinical features of dysregulation of the innate and adaptive immune system. Lastly, NETosis (neutrophils forming extracellular traps to immobilize and kill invading microbes) may play a role in BS. By netting, Neutrophils externalize autoantigens, making them more exposed to the adaptive and innate immune system. The role of NETosis in BS has not been assessed yet.

Study objective

Main objective is to collect longitudinal follow up data of all patients with (suspected) Behçet*s syndrome. In order to study prevalance and incidence of clinical disease manifestations.
Secondary objectives:
1. What is the prevalence of various disease manifestations (for example uveitis and DVT) and damage accumulation (like visual loss and cataract) in patients with BS in the Netherlands?
2. What is the level of disease activity in patients with Behçet*s syndrome as well in those with suspected BS, when measured using the BD-CAF, BSAS and ARMD BD-HAQ?
3. What is the quality of life in patients with (suspected) BS, does it correlate with disease activity or damage accumulation?
4. What is the incidence of pathergy positivity? Is patient evaluation of pathergy testing equally reliable, compared to a) real time assessment by a dermatologist or b) assessment of photographs by a trained physician?
5. To study genetics in patients with BS (in relation to clinical manifestations)
6. To create a prediction rule for patients with suspected BS.
7. To study immune regulation in patients with (suspected) BS, specifically clonal T cell expansion and antigen presentation via MHC1.
8. To study the role of NETosis in patients with (suspected) BS.

Study design

observational, longitudinal.
biobanking

Study burden and risks

The burden of our first studyvisit (for entirely NEW patients) consists of the collection of 2 tubes of blood for regular care purposes and 14 to a maximum of 15 additional tubes of blood for study purposes. Also, patients are asked to complete 3 additional questionnaires, of which once a Quality of Life questionnaire.
For those patients who come for visit 2 (were included in the former cross sectional Behcet study allready), 14 extra tubes of blood will be drawn. And 2 extra questionnaires (not the QoL questionnaire).

Regarding follow up:
In patients who consent for the follow up study protocol (those who were included in the cross sectional study earlier and the new patients as well), all information of all follow up visits will be added to the database, with 2 additional questionnaires, the type of follow up will be either yearly or every visit information will be put in the database. Every follow up visit 2 extra questionaires will be done. Once a year 3 extra tubes of blood will be drawn (all patients).

In case of a flare 14 extra tubes of blood and 2 questionnaires (this is once).

Benefit: more understanding of the disease, better care for individual patients due to the dedicated nature and extensive typing of complaints. Specifically for those with a probable BS the benefit will be creating a prediction model.

Contacts

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Trial sites

Listed location countries

Netherlands

Eligibility criteria

Age
Adults (18-64 years)

Elderly (65 years and older)

Inclusion criteria

Adult patients who fullfill the international criteria of Behcet syndrome.
Those with a clinical suspicion of Behcet syndrome.

Exclusion criteria

none (except no informed consent)

Study design

Design

Study type: Observational invasive

Masking:	Open (masking not used)
Control:	Uncontrolled
Primary purpose:	Basic science

Recruitment

NL

Recruitment status:	Recruiting
Start date (anticipated):	03-02-2017
Enrollment:	200
Type:	Actual

Ethics review

Approved WMO Date:	15-09-2016
Application type:	First submission
Review commission:	METC Amsterdam UMC
Approved WMO Date:	07-07-2017

Application type:	Amendment
Review commission:	METC Amsterdam UMC

Study registrations

Followed up by the following (possibly more current) registration

No registrations found.

Other (possibly less up-to-date) registrations in this register

No registrations found.

In other registers

Register	ID
CCMO	NL58955.048.16